

# Striving for a World Without Primary Sclerosing Cholangitis

Compassionate Allowance  
Presentation: Primary  
Sclerosing Cholangitis  
PSC Partners Seeking a Cure  
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# PSC Partners Seeking a Cure – Organization Overview



- Founded in 2005
- 501(c)(3) nonprofit organization

The mission of PSC Partners Seeking a Cure is to provide education and support to PSC patients, families and caregivers and to raise funds to research causes, treatments and a cure for primary sclerosing cholangitis.

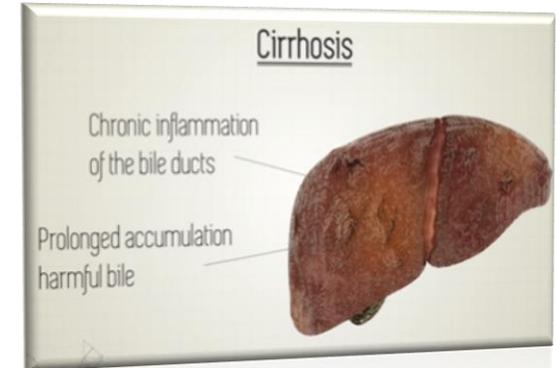
# PSC Partners Seeking a Cure – Programs



- Annual conferences for patients and caregivers
- Research grant program – over \$2.6 million awarded to date
- Patient registry – over 1,100 patients enrolled



# Primary Sclerosing Cholangitis – Basic Overview



- Chronic, progressive, and rare disease affecting the bile ducts inside and outside of the liver
- Bile ducts become inflamed, leading to scarring and narrowing of the ducts, blocking the flow of bile
- The liver is damaged by trapped bile, often causing fibrosis and cirrhosis of the liver, leading to liver failure

# Primary Sclerosing Cholangitis – Basic Overview, Continued



- PSC is commonly undiagnosed or misdiagnosed
- Individual disease progression is unpredictable
- Affects over 30,000 patients in the U.S. and has a 2:1 male: female ratio
- Over 75% of PSC patients also have inflammatory bowel disease (IBD), most commonly ulcerative colitis
- Commonly coincides with other autoimmune diseases





# Primary Sclerosing Cholangitis – Symptoms

- Pruritus or intense itching
- Extreme fatigue
- Abdominal pain
- Jaundice

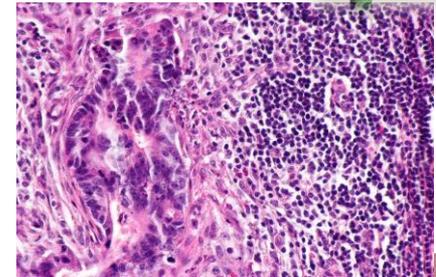
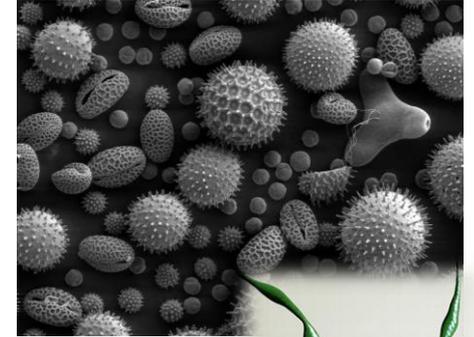


# Primary Sclerosing Cholangitis – complications



Complications from PSC can be life-threatening, and include:

- Bacterial cholangitis
- Biliary stone disease
- Dominant stricture (especially in the common bile duct)
- Cholangiocarcinoma (cancer of the bile ducts)
- Heightened risk of colon cancer for patients with IBD
- Hepatic encephalopathy
- Varices
- Ascites
- Bleeding disorders



# Primary Sclerosing Cholangitis – Cause and Treatments



- No known cause
- No treatment to slow progression
- Current treatments aim to relieve symptoms
- Only definitive treatment is liver transplantation
- Even following transplant, PSC can recur in the new liver



# Primary Sclerosing Cholangitis – Onset, Methods of Diagnosis



- PSC mostly affects middle-aged men, but also affects children and women
- Onset can occur at any age
- Methods of diagnosis:
  - Imaging and blood tests
  - Liver biopsy



# Primary Sclerosing Cholangitis – Disability



- PSC is recognized as a chronic liver disease at 5.00D2 (adults) and 105.00D2 (children)
- As PSC progresses, symptoms become increasingly debilitating and severely diminish the patient's quality of life
- PSC is coded separately in ICD-11, and is currently coded under K83.0 "Cholangitis" in ICD-10 CM. Request for separate ICD-10 CM code is currently pending
- Most PSC patients have more than one autoimmune disease, which severely compromises the patient's ability to engage in meaningful work.

# Recommendations for SSA



- Include primary sclerosing cholangitis in the list of conditions eligible for Compassionate Allowance

# Thank You!

